# An Atypical Case of Foster Kennedy Syndrome

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***In memoriam*** to Pierre Lasjaunias.

## Summary

*Foster-Kennedy syndrome was described in 1911 as an ophthalmologic manifestation of compression by a solid tumor in the frontal area with intracranial hypertension (ICHT). We describe a peculiar case of Foster-Kennedy syndrome associated with an arteriovenous malformation in which neither optic nerve compression nor ICHT was obvious. We discuss the different pathogenic mechanisms to explain this case, for which a chronic venous hypertension was the most probabile etiology.*

## Introduction

In 1911, Foster Kennedy described a series of patients with a frontal intracranial expanding process associated with ipsilateral optic atrophy, controlateral papilledema and anosmia[1]. Almost all cases were caused by a solid intracranial tumor, usually a meningioma of the basal frontal lobe, the olfactory groove, or the medial third sphenoid wing [2,3]. Classically, Foster Kennedy syndrome is due to a direct compression of the ipsilateral optic nerve by the expanding process leading to optic atrophy, while the swelling controlateral optic disc reflects intracranial hypertension (ICHT) secondary to the tumor's mass effect. We describe a peculiar case of arteriovenous malformation (AVM) inducing a Foster Kennedy-like syndrome, in which neither optic nerve compression nor ICHT was obvious.

## Case Report

A 33-year-old man was referred for a right frontal AVM, initially revealed by headaches, and treated by four sessions of histoacryl transarterial embolization. On arrival, visual acuity was 20/22 in the left eye, and no light perception in the right eye. Slit lamp examination of both eyes was normal, with the exception of an edematous left optic disc and an atrophic right optic disc (Figure 1). Complete neurological assessment was normal. No headache, nausea, vomit or diplopia was reported. Goldman visual field of the left eye revealed a nasal deficit with an excluded blind spot (Figure 2). Cerebral resonance magnetic imaging (RMI) showed an AVM localized in the interior and anterior frontal lobe, distant from the optic nerves and from the base of the skull. There was no radiological sign of ICHT (Figure 3). Cerebral angiography showed that the AVM was located in the frontal interior circumvolution, principally occupying the gyrus, and was fed by branches of the anterior frontal artery. The drainage was exclusively cortical to the superior longitudinal sinus. There was a pial reflux in the frontal and parietal areas, suggesting a venous hypertension into the sinus.

Transarterial embolizations resulted in a significant decrease of the arteriovenous shunt (nearly 70% as showed by angiography, (Figure 4)), but the AVM was not completely excluded. Owing to the optic disc edema, the patient was treated with high dose steroids (intravenous methylprednisolone 500 mg QD for three days) and oral acetazolamide (250 mg BID). One month later, the disc edema has significantly reduced, and visual acuity was stable.

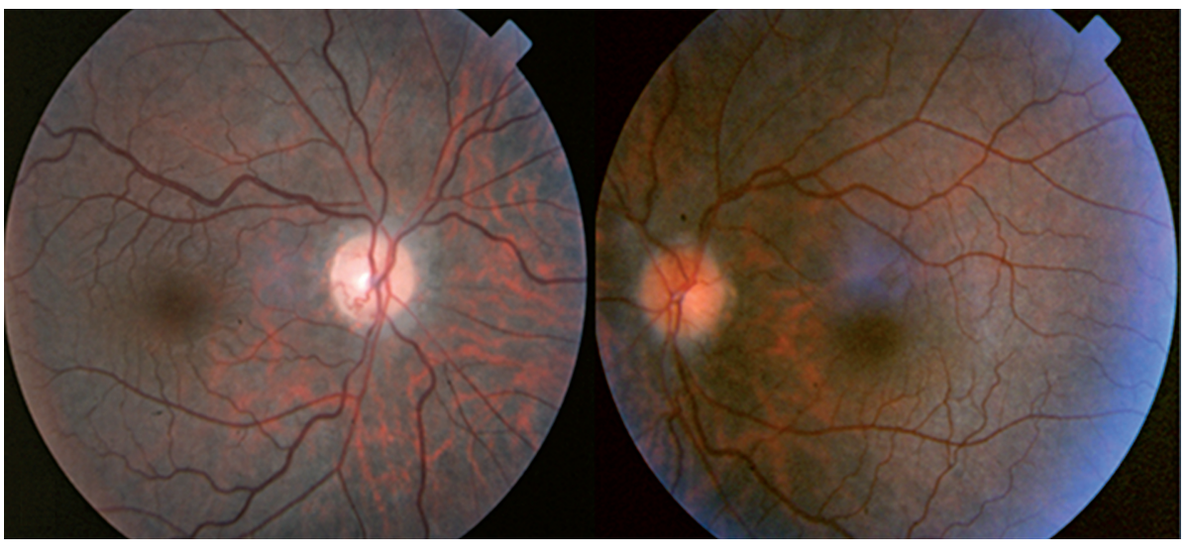


*Figure 1* Atrophy of the right optic disc (A). Oedema of the left optic disc (B).

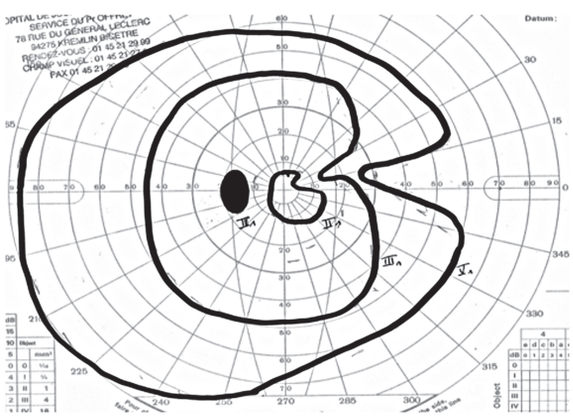


*Figure 2* Goldman visual field of the left eye showing a nasal deficit with an excluded blind spot. Visual field of the right eye was not possible to assess (no light perception).

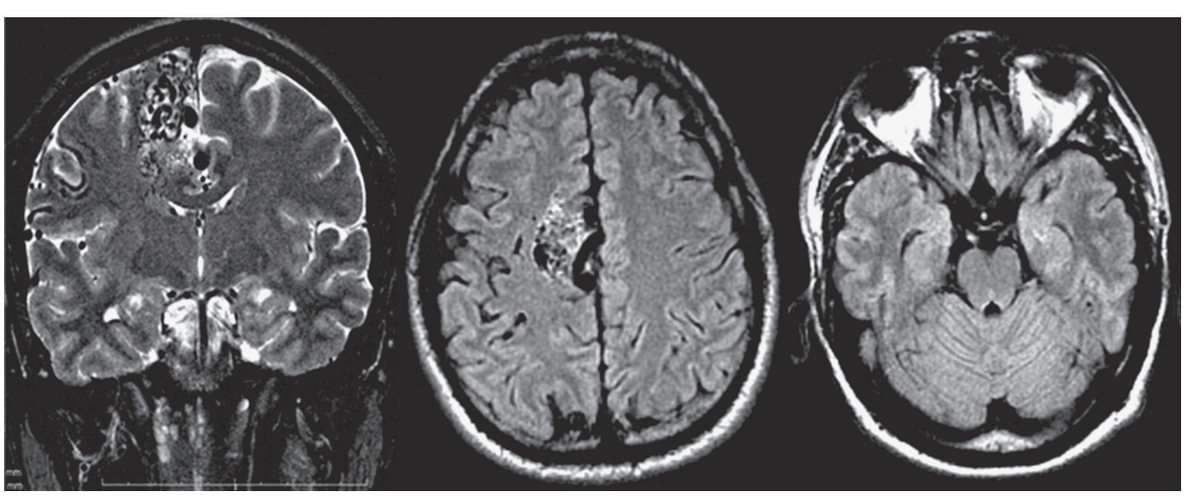


*Figure 3* Cerebral magnetic resonance imaging: no radiological sign of intracranial hypertension and no lesion in optic pathways (A: frontal section; B,C: axial section).

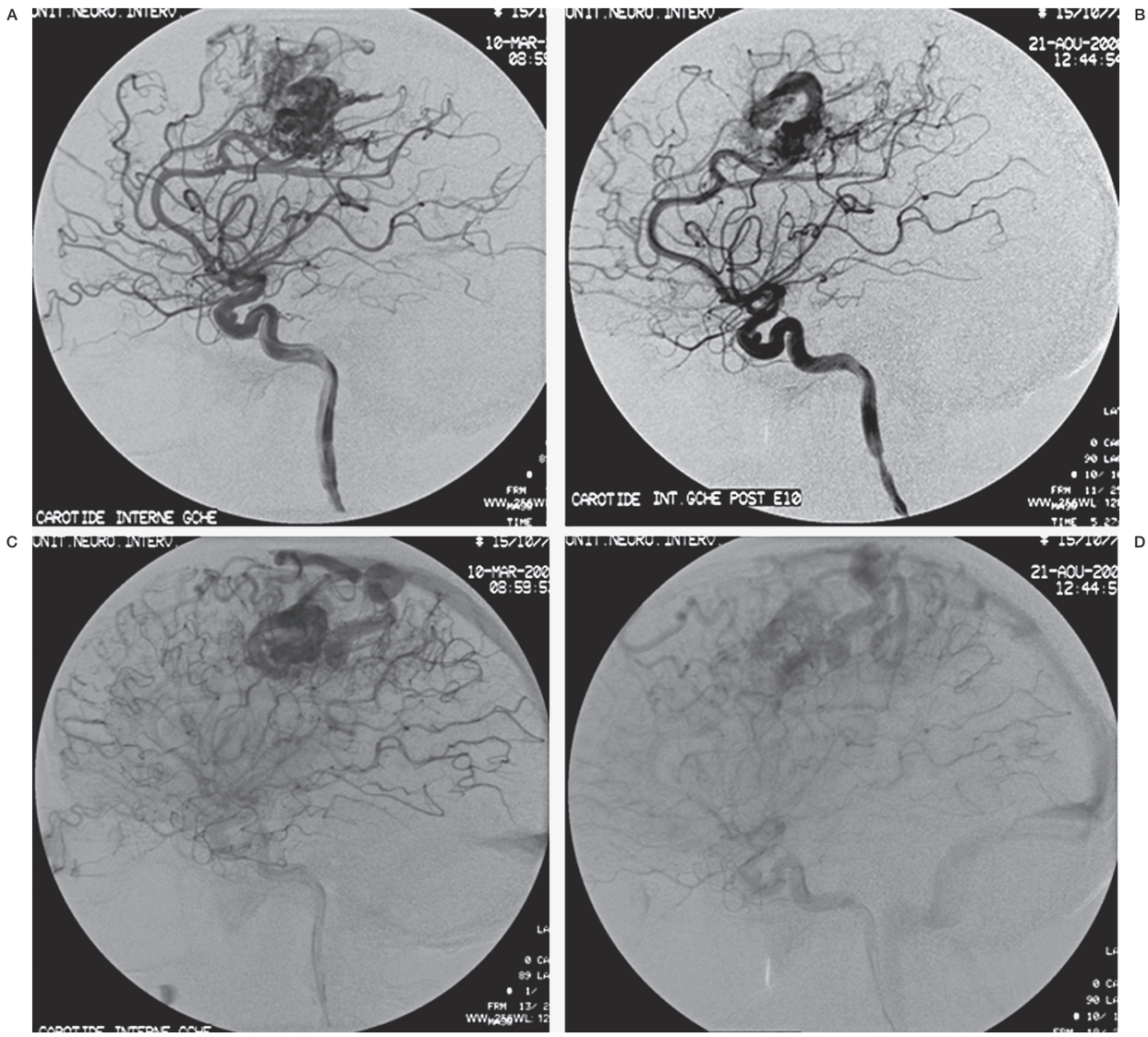


*Figure 4* Cerebral angiography before (A) and after (B) embolization showing a significant reduction of the shunt related to arteriovenous malformation. Lateral views depicting venous phase at selective digital subtraction angiography of the left internal carotid artery before (C) and after staged embolization of the right frontal AVM (D): varicosity of the cortical draining veins the malformation is reduced after endovascular treatment reflecting lessened venous pressure.

## Discussion

Foster-Kennedy syndrome is a rare condition. A review of literature shows that despite a common pattern in ophthalmologic signs, all FKS do not share the complete pathogenic settings. Three types of FKS could thus be distinguished: type 1 with unilateral and direct optic nerve compression with ICHT[1], type 2 with bilateral and direct optic nerve compression but no ICHT [4], and type 3 with chronic ICHT but no nerve compression [4]. Type 1 is the classical presentation as defined above. In FKS type 2, the bilateral and asymmetric compression of both optic nerves by the tumor explains the atrophy for the most compressed optic nerve (long-term compression) and the papilledema in the other (short-term compression) [4]. In FKS type 3, no direct compression by the tumor is observed; and optic disc atrophy reflects the end-course of an asymmetrical chronic papilledema due to chronic ICHT [4]. As visual progno-

sis depends on the duration of papilledema (an atrophic optic nerve is definitively injured), a symptomatic treatment using steroids, to reduce inflammation and edema, and carbonic anhydrase inhibitors, to reduce ICHT, may accelerate vision recovery, but etiologic treatment remains mandatory [6].

In our patient, a Foster-Kennedy-like syndrome was observed despite no obvious optic compression, and no ICHT. The AVM was not in contact with the optic pathways or with the base of the skull, and there was no mass effect or ICHT. It is thus likely that chronic venous congestion in the frontal area induced by the MAV may have played an important role in the onset of the bilateral optic nerve disease [5]. The management of AVM depends on vascular architecture, which rules the transarterial embolization. Due to the relative completeness of embolization and the satisfying clinical evolution, no additional neurosurgery and/or radiosurgery was used in our patient.

To the best of our knowledge, this is the first case of FKS related to an AVM with a chronic venous congestion and ischemia in the frontal area, rather than a direct compression and/or an intracranial hypertension. Besides transarterial embolizations, medical treatment succeeded in reducing papilledema, preventing a decrease of visual acuity.

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